Clinical Guideline

FONTAN CIRCULATION (TOTAL CAVOPULMONARY CIRCULATION (TCPC))

SETTING	South West England and South Wales
GUIDELINE FOR	Cardiology teams in South West England and South Wales hospitals
PATIENT GROUP	Adult patients with congenital heart disease

GUIDANCE

Follow-up: annual
Associated lesions: dependent on the underlying abnormality, note may be isomerism
Inheritance: dependent on the underlying abnormality

Long-term complications:

1. Deterioration of ventricular function
2. AV valve regurgitation
3. Fontan obstruction ('peel', Dacron>GoreTex)
4. Right pulmonary venous obstruction -by enlarged RA in patients with RA-PA Fontan
5. Bradyarrhythmia -sinus node dysfunction and heart block – unmask during exercise testing, pace via coronary sinus or surgically
6. Atrial tachyarrhythmia -affects > 50% with A-P Fontan vs. 10-20% of patients with lateral tunnel/TCPC; can result in profound haemodynamic compromise/clot formation. Aim to restore sinus rhythm. Often resistant to drugs/EP.
7. Thromboembolism -systemic and pulmonary, may be associated with AF, slow flow and clotting abnormalities (e.g. protein C deficiency). RA clot especially common in atriopulmonary Fontan.
8. Fontan associated liver disease (FALD) -congestion, dysfunction, cirrhosis, hepatocellular carcinoma (AFP may be normal) and varices
9. PLE -in 10%, poor prognostic sign - 5-year survival 50%. If obstruction, may be cured my relieving this. Associated with ascites, peripheral oedema, pleural and pericardial effusions, chronic diarrhoea and elevated stool α1 antitrypsin levels with low serum albumin. Lymphatic interventions emerging therapy.
10. Plastic bronchitis - very poor prognostic sign
11. Cyanosis -due to fenestration, veno-venous collaterals draining to pulmonary veins/ LA/ conduit dehiscence or development of pulmonary AVMs.
12. Narrowing of the proximal descending aorta (post Norwood)

Annually:
History:
reducing exercise capacity/fatigue
dyspnoea
palpitations
syncope
haemoptysis/bleeding

	diarrhoea/recurrent infections think PLE weight loss (think HCC)
Exam:	may depend on original anatomy non-pulsatile and high JVP should be quiet on auscultation single S2 PSM if AV valve regurgitation, EDM if AR absent or weak radial pulse post BT shunt hepatomegaly (think obstruction/high pressure, if Glenn this may be the only sign as JVP will not be up) signs of chronic liver disease abdominal distension (think HCC) sats – compare with previous if ascites, peripheral oedema, pleural effusions (look for PLE)
ECG:	dependent on original anatomy confirm sinus rhythm (compare to old ECG- may be asymptomatic slow IART. Junctional rhythm common)
Echo:	Biphasic flow (increases with inspiration) in SVC and hepatic veins (vmax should be <1.5m/s) systemic ventricular function- TVI values, M-Mode, and strain AV valve regurgitation aortic root and valve thrombus in right atrium fenestration on CFM pulmonary venous return-assess for a gradient in visible veins
Bloods:	FBC, clotting, U+E, ferritin, LFTs (to include AST and γGT), BNP, AFP, serum protein and albumin. If low albumin, stool α 1 anti-trypsin for PLE. Baseline viral hepatitis and hemochromatosis screen (once only)
Liver imaging:	annual USS for cirrhosis/masses/ spleen size, portal HT Fibroscan – 3 yearly (email gastrohepsecs@uhbw.nhs.uk) if endoscopy/MRI/CT as directed by hepatology (Dr James Orr)
Further investigations:	
CXR:	not routine. Normal heart size and pulmonary vascularity. May see calcification of TCPC. If pleural effusions, search for PLE.
CPET:	baseline and routinely every 5 years-otherwise if symptoms change or if considering transplant
Holter:	if palpitations, pre-syncope or syncope
TOE:	to assess AV valve regurgitation further for potential surgery
Catheter:	if well, consider every 10 years if new symptoms, ventricular dysfunction, arrhythmias, cyanosis or suspected obstruction

	to assess haemodynamics/ obstruction and cause for worsening cyanosis (AVMs, collaterals) creation of a fenestration may be needed to decrease Fontan pressure
EP study:	if documented atrial arrhythmias. If pacing required- to discuss.
MRI:	at baseline to confirm anatomy and assess function/patency of Fontan pathway, collaterals and pulmonary vein obstruction by enlarged RA, thrombus, cardiac output repeat if change in symptoms or ventricular function. If well, every 3-5 years.
CT:	if suspected thrombus in Fontan or to visualise existing stents or if MRI not possible due to pacemaker.
Drugs:	Anti-coagulate. No robust data on NOACs but increasing use. ACE/ARB reasonable if systemic ventricular dysfunction If right atrial isomerism need to be on penicillin and receiving annual pneumovax no hard evidence for pulmonary vasodilators
Pregnancy:	Relatively low risk to woman if uncomplicated Fontan. Avoid if any complication. High risk of miscarriage/severe IUGR/prematurity/fetal death (up to 60%). Pre-pregnancy counselling mandatory.
Contraception:	not for COCP/oestrogen containing preparations
Endocarditis:	antibiotic prophylaxis before high-risk dental work for all
Exercise:	moderate symptom-limited aerobic exercise recommended

Discuss if:

- New ventricular dysfunction/heart failure
- Worsening exercise capacity/cyanosis
- Tachy- or bradyarrhythmias (Fontan conversion may be considered if resistant tachyarrhythmia)
- ≥Moderate AV valve or aortic regurgitation
- Fontan obstruction
- Sub-aortic obstruction
- Progressive aortic root dilatation/narrowing with hypertension
- Pulmonary venous obstruction
- PLE (admission, IV Furosemide, albumin solution, sc heparin, spironolactone, prednisolone/budesonide, ACEI, dietitian consultation for high protein low fat diet and salt restriction)

Refer to hepatology if LFTs or liver USS abnormal

Appendix 1 – Evidence of Learning from Incidents

The following table sets out any incidents/ cases which informed either the creation of this document or from which changes to the existing version have been made.

Incidents	Summary of Learning
n/a	

Table A

REFERENCES	Baumgartner H, De Backer J, Babu-Narayan SV, Budts W, Chessa M, Diller GP, Lung B, Kluin J, Lang IM, Meijboom F, Moons P. 2020 ESC Guidelines for the management of adult congenital heart disease. Eur Heart J. 2020. Stout et al. 2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease. Journal of the American College of Cardiology Aug 2018, 25255; DOI: 10.1016/j.jacc.2018.08.1029 Rychik J, Atz AM, Celermajer DS, Deal BJ, Gatzoulis MA, Gewillig MH, Hsia TY, Hsu DT, Kovacs AH, McCrindle BW, Newburger JW, Pike NA, Rodefeld M, Rosenthal DN, Schumacher KR, Marino BS, Stout K, Veldtman G, Younoszai AK, d'Udekem Y; American Heart Association Council on Cardiovascular Disease in the Young and Council on Cardiovascular and Stroke Nursing. Evaluation and Management of the Child and Adult With Fontan Circulation: A Scientific Statement From the American Heart Association. Circulation. 2019 Jul 1. Greenway SC, Crossland DS, Hudson M, Martin SR, Myers RP, Prieur T, et al. Fontan-associated liver disease: Implications for heart transplantation. The Journal of Heart and Lung Transplantation 2016;35:26-33.
RELATED DOCUMENTS AND PAGES	Regional Referral Guidance for Adult Patients with Congenital Heart Disease RegionalReferralGuidanceAdultPatientsWithCongenita-3.pdf Regional Referral Pathway for Cardiac Disease in Pregnancy ClinicalGuidelineForCardiacDiseasePreExistingOrPre-1.pdf
AUTHORISING BODY	Cardiac Executive Group, Bristol Heart Institute
SAFETY	None
QUERIES AND CONTACT	**Bristol:** Contact any of the following via UHBW switchboard – 0117 923 0000 Dr S Curtis Dr G Szantho Dr M Turner Dr R Bedair ACHD Specialist Nurse Team 0117 342 6599 **Cardiff:** via UHWales switchboard - 029 2074 7747 Dr S MacDonald Dr H Wallis Dr DG Wilson Dr N Masani ACHD Specialist Nurse Team 02920 744 580
AUDIT REQUIREMENTS	Adherence to guideline will be audited periodically as part of ACHD departmental audit

Plan Elements	Plan Details
The Dissemination Lead is:	Dr Stephanie Curtis
Is this document: A – replacing the same titled, expired SOP, B – replacing an alternative SOP, C – a new SOP:	A
If answer above is B: Alternative documentation this SOP will replace (if applicable):	
This document is to be disseminated to:	South West and South Wales Congenital Heart Network
Method of dissemination:	Email
Is Training required:	No

Document Change Control

Date of Version	Version Number	Lead for Revisions	Type of Revision	Description of Revision
Jan 2021	2	Consultant Cardiologist	Minor	Updated contacts and related documents. Under complications: “May need Fontan conversion and MAZE” has been removed. “Lymphatic interventions emerging therapy” added. Under bloods LFTs: “to include AST” added. Under liver imaging: ”Fibroscan – 3 yearly (email gastrohepsecs@uhbw.nhs.uk)” added. Under catheter ‘consider’ every 10 years added. Under CT: “to visualise existing stents” added Under drugs NOACS “but increasing use” added Endocarditis prophylaxis recommended for all.